Systemic Lupus Erythematosus among Jordanians: A Single Rheumatology Unit Experience

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ABSTRACT

Objectives: To study the characteristics of Systemic Lupus Erythematosus in Jordan and compare results with other Arab countries in the region.

Methods: This descriptive study was conducted on 50 Jordanian patients with Systemic Lupus Erythematosus attending the rheumatology clinic at King Hussein Medical Center between 2000 and 2007. King Hussein Medical Centre is one of the main referral hospitals in Jordan. Data was collected from patients, relating to the various disease manifestations and mode of presentation, age at disease onset, disease duration, and family history of Systemic Lupus Erythematosus. Statistical analysis was performed using simple descriptive and bivariate statistics, such as mean, median, range and Chi square.

Results: Forty-four females and six male patients were included. Female to male ratio was 7.3:1 with median age of 22 years (range 14-48). Mean disease duration was 43 months (range 12-90 months). Clinical manifestations of Systemic Lupus Erythematosus in descending order were, musculoskeletal (90%), cutaneous and oral ulcers (85%), renal complications (50%), neuropsychiatric (45%), haematological (40%), pulmonary complications (30%), and cardiac complications in 15% of cases. Secondary anti-phospholipid antibody syndrome (APS) occurred in 30% of cases. Three patients died from severe chest infection. Another patient died from massive sagittal sinus thrombosis and severe hydrocephalus. A comparison between our findings and published Systemic Lupus Erythematosus studies from four Arab countries revealed no significant clinical differences.

Conclusion: Systemic Lupus Erythematosus is an important rheumatological disease with serious consequences including morbidity and mortality. It behaves in a similar way to Systemic Lupus Erythematosus in other Arab countries. Renal, pulmonary and neurological complications are the main cause of morbidity. Chest infection is a major cause of mortality.

Key words: SLE, Vasculitis, Jordan, Arabs

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Introduction

Systemic Lupus Erythematosus (SLE) is a chronic inflammatory autoimmune disease of unknown aetiology, which is recognized worldwide. Many observations suggest a role for genetic predisposition, hormonal, immunological and environmental factors in its pathogenesis.[1] It is a multi-system disease that produces symptoms varying from mild to life threatening. Marked variations in the frequencies of SLE amongst differing racial groups have been noticed.[2] Studies from Arab countries are few.[3-8] In this study, we attempt to analyze SLE characteristics in Jordanian population and compare it with similar studies in other Arab countries.

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This study was conducted to study the characteristics of Systemic Lupus Erythematosus in Jordan and compare results with other Arab countries in the region.

Methods

This is a descriptive study to involving patients with SLE attending a rheumatology clinic at King Hussein Medical Center (KHMC) between 2000 and 2007. Fifty Jordanian patients were included in the study, all fulfilling the 1982 ACR revised criteria for the diagnosis of SLE and followed up by same attending rheumatologist.[3]

Data collected from patients, relating to the various disease manifestations and mode of presentation, age at disease onset, disease duration, and family history of SLE. All patients had full CBC, ESR, ANA, dsDNA, ENA, anti-cardiolipin antibodies, RF, CRP, a-PTT, lupus anticoagulant and x-ray of chest, hands and feet.

Statistical analysis was performed using simple descriptive and statistics, such as mean, median and range and Chi square.

Results

Fifty patients with SLE were studied, 44 females and six males with a ratio of 7.3:1. All patients were Jordanians from Arab descent. The median age of patients was 22 years (range 14-48 years). The mean duration of follow up was 32 months (range 3-84 months).

The females: male median age of disease onset was 20.5: 21 years respectively. The range of disease duration was 43 months. Three patients (6%) had a family history of SLE. Demographic data is shown in Table I.

The different clinical manifestations among the study group from Jordanian and other Arabic countries are summarized in Table II. Forty patients (80%) presented with skin lesions suggestive of SLE, alopecia (60%), butterfly rash (50%), painless mouth ulcers (30%), photosensitivity (20%), and discoid lupus (8%).

Musculoskeletal manifestations are the commonest presenting symptoms, and occurred in 90% of patients. Arthralgia was noticed in 80% of cases, non-erosive arthritis in 60%, and myalgia in 40% of cases. Aseptic necrosis of the hip was observed in four (8%) patients proved by Magnetic Resonance Imaging (MRI).

Twenty five patients, (23 females and 2 males) had evidence of renal involvement ranging from mild to severe proteinuria, abnormal urinary casts and raised serum creatinine. Twenty patients had proteinuria more than one gram per 24 hours underwent kidney biopsy. According to WHO classification of renal glomerulonephritis, two patients had moderate mesangial lupus nephritis (class II), five patients had focal segmental proliferative lupus nephritis (class III), ten patients had diffused proliferative lupus nephritis (class IV) and three patients had membranous glomerulonephritis (class V nephritis).

Two female patients needed temporary haemodialysis, both responded well to steroid and immunosuppressive therapy. Another two patients went into end stage renal disease and needed long-term renal replacement therapy. There was no difference in the occurrence of renal complications between both sexes though the number was small to make statistical conclusion.

Many patients presented with central nervous system involvement. Thirteen patients (26%) presented with different neuropsychiatric complications of SLE. Six patients (12%) presented with psychiatric symptoms ranging from mild depression to severe psychosis. Seizures were observed in three patients (6%), trigeminal neuralgia (4%), stroke (4%), and aseptic meningitis (4%).

Forty percent of patients had evidence of hematological disorder. Leucopenia was the commonest haematological finding observed in 35% of patients. Anemia was seen in 26% of patients, 12% of these patients had evidence of hemolytic anemia. Thrombocytopenia was noticed in 20% of patients.

Fifteen patients (30%), 14 females and one male patient had evidence of APS according to antiphospholipid syndrome classification criteria [4]. Another two patients, one female and one male had deep vein thrombosis without evidence of antiphospholipid syndrome. There was significant association between APS and deep vein thrombosis ($P \leq 0.05$). Ten patients (20%) presented with deep vein thrombosis and five (10%) patients with recurrent deep vein thrombosis. Five patients (10%) developed sagittal sinus thrombosis. Two patients had axillary vein thrombosis and another patient developed hepatic vein thrombosis.

Five patients (30%) had positive lupus anticoagulant, six patients (33%) had positive anticardiolipin antibodies and six patients (33%) had

Table I. Demographic characteristics of 50 Jordanian patients with SLE

	No of patients	%
Female	44	90
Male	6	10
Female: male ratio	7.3:1	
Mean age of disease onset		
Female	21.9	
Male	22.3	
Family history	3	6

Table II. Clinical and serological characteristics of 50 SLE patients seen at King Hussein Medical Centre compared with SLE patients from other Arab countries

Country	Jordan	Lebanon[4]	Kuwait[5]	UAE[6]	SA[7]
No. of patients	50	100	108	33	87
F:M ratio	7.3:1	6.1:1	9.8:1	15.1:1	8.7:1
Median age	23	25	31.5	26	28.5
Clinical features%					
(ACR 1982 revised criteria)					
Malar rash	60	52	43	36	56
Discoid lupus	16	19	10	3	18
Photosensitivity	25	16	48	42	26
Oral ulcers	50	40	33	27	16
Arthritis	80	95	87	91	91
Serositis	26	40	29	33	56
Renal	50	50	37	63	63
Neuropsychiatric	20	10	23	9	25
Hematological	40	47	53	45	78
Leucopenia	35	17	83	30	33
Anaemia	28	-	-	-	-
Hemolytic anemia	12	10	-	9	-
Thrombocytopenia	20	33	26	21	21
Positive dsDNA	85	50	58	97	93
Antiphospholipid syndrome	30	5	21	33	10
Positive ANA	95	87	94	89.5	98

both tests positive. Seven patients (50%) presented with recurrent abortions. Six patients (12%) died during the study period in spite of all treatment modalities. Three patients died of severe chest infection. Two patients died of severe haemorrhagic pneumonitis. One patient died of severe hydrocephalus secondary to massive sagittal sinus thrombosis.

Discussion

In this study, the clinical spectrum of a group of Jordanian patients with SLE was presented and results were compared with similar published studies in other Arab countries.

The female to male ratio is 7.3:1, which is in agreement with published reports in Lebanon and Saudi Arabia and less than what was observed in Kuwait and United Arab Emirate.[5-8] The study confirms that it is a disease of young people. The median age of the disease onset is the third decade, and there is no significant difference between ages of disease onset in both sexes. These results are similar to published studies in the region.

The clinical spectrum of the disease was almost similar to the clinical manifestation of the disease in other countries in the regions. Musculoskeletal involvement was the commonest presenting feature of the disease (80%) and is comparable to those published reports in the region. The cutaneous features are the second commonest presenting feature (60%). Renal involvement was the main cause of increased morbidity. Lupus nephritis affected half of the patients (50%) which is almost similar to the published studies in UAE (54%), Lebanon (50%), but less than those reported in Saudi Arabia (63%) and more than those reported in

Kuwait (37%). Three patients went into end stage renal disease and needed renal replacement therapy. The reason for the high rate of renal complications in our cohort of patients is that we have easy access and direct referral system to our clinics and good collaboration with hospital renal unit.

Secondary antiphospholipid syndrome has a common association with SLE. It is an autoimmune disease which is characterized by antiphospholipid antibodies and one clinical manifestation, the most common being venous or arterial thrombosis and recurrent fetal loss.[4] Love and Santro in their analysis of over 1000 patients with SLE found an average prevalence of 34% of lupus anticoagulant and 44% for anticardiolipin antibodies.[11] Secondary lupus anticoagulant constituted 30% of our group of patients, which is significantly higher than previously reported studies in Lebanon (5%), Kuwait (20%) and Saudi Arabia (10%). The high rate of APS in our group of patients compared to other Arab countries in the region might be that King Hussein Medical Centre is the main referral center in Jordan and the difficult and complicated cases are more likely to be referred to it.

Vascular complications are an important cause of increased morbidity. Ten patients presented with deep vein thrombosis including, five patients with recurrent deep vein thrombosis, five patients presented with sagittal sinus thrombosis. Unfortunately, there are no published data regarding vascular complication of the disease to compare with. Haematological complications of the disease are almost similar to results in other Arab countries.

SLE could be a serious disorder despite of all available treatment options, though the mortality rate has improved remarkably since the introduction of steroids and other immunosuppressant drugs. The leading causes of mortality in SLE are irreversible renal damage, fulminant infection and uncontrolled disease activity.[12,13]

Mortality rate in our study was 12%, which is in concordance with other published international studies, though there is no mortality data published in the region to compare with. The main cause of mortality in our study was severe chest infection, then haemorrhagic pneumonitis and massive sagittal sinus thrombosis. The spectrum of infection was similar to that published in most major studies.[14]

Renal involvement was an important cause of increased morbidity, however, this did not add to increase mortality in our group of patients. This could be attributed to the aggressive management and availability of renal replacement therapy. Similar observation was noticed by Al-Jarallah in Kuwait.[5]

Conclusion

SLE in Jordan behaves in a similar way to other Arab countries in the region. Chest infections and haemorrhagic pneumonitis are the main cause of mortality. Secondary antiphospholipid syndrome is a common associated complication.

Further epidemiological studies are needed to find out the exact incidence and prevalence of SLE in Jordan.

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